Indian J. Otolaryngol. Head Neck Surg.
(July-September 2007) 59:273–276

Case Report

Primary extraskeletal Ewing's sarcoma of the maxilla with intraorbital extension

Saurabh Varshney · S. S. Bist · Nitin Gupta · Rajat Bhatia

Abstract

Extraskeletal Ewing's sarcoma is often described as a tumour involving the soft tissues of the lower extremities and the paravertebral region. Primary Ewing's sarcoma of the cranium is extremely rare, with only 17 cases reported so far [24]. Involvement of the paranasal sinus is a very rare entity. Involvement of facial bones is characterized by clinical and radiological features distinct from those commonly observed in other sites. Because of the above peculiarities a delay in diagnosis and thus in starting treatment is very probable in such cases. Primary Ewings sarcoma rarely arises in the facial skeleton and only occasionally in the maxilla. To date, there have been 22 cases of maxillary Ewing's sarcoma reported in the English-language literature [25]. We report here a new case of Ewing's sarcoma localized to the maxillary sinus, nose and the orbit, successfully treated by surgery, local high dosage radiotherapy and systemic chemotherapy.

Keywords Ewing's sarcoma · Paranasal sinus

S. Varshney[1] (✉) · S. S.Bist[2] · N. Gupta[3] · R. Bhatia[3]
[1]Professor & Head
[2]Assistant Professor
[3]Senior Resident
Department of E.N.T.
Himalayan Institute of Medical Sciences
Jollygrant; Doiwala
DEHRADUN - 248 140
(Uttaranchal)
India

e-mail: drsaurabh68@gmail.com

Ewing's sarcoma is a malignant neoplasm of uncertain histogenesis, most commonly arising in the skeleton. The head and neck are unusual sites for primary Ewing's sarcoma. Primary Ewing's sarcoma of the paranasal sinus (PNS) is exceptionally rare and most cases are described in the maxillary sinus [16]. Of the 14 cases that we are aware of reported in the literature, the maxillary sinus (7 cases) is the most common site followed by the ethmoid sinus (2 cases) and the nasal fossa (5 cases) [1–10].

Case report

An adolescent 15-year-old male, who had polio in childhood was admitted in the ENT ward, Himalayan Institute of Medical Sciences, Dehradun, India, with the complaints of left nasal obstruction, nasal discharge, decreased vision, watering from left eye, protrusion of left eye associated

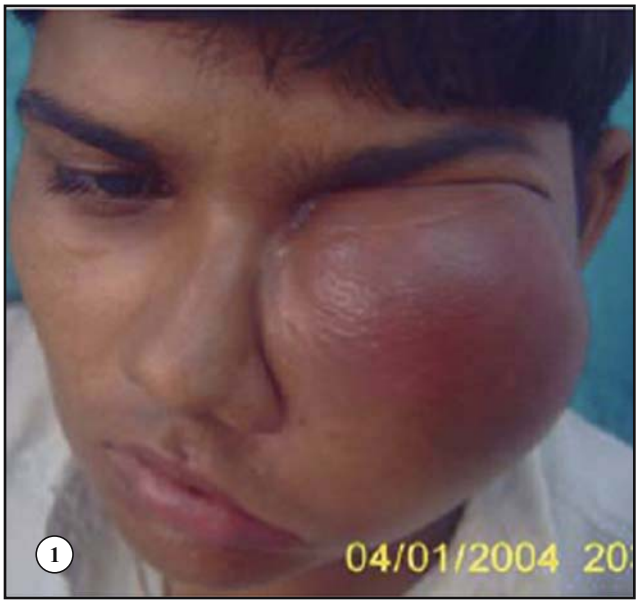


Fig. 1 Clinical photograph

Indian J. Otolaryngol. Head Neck Surg.
(July-September 2007) 59:273–276

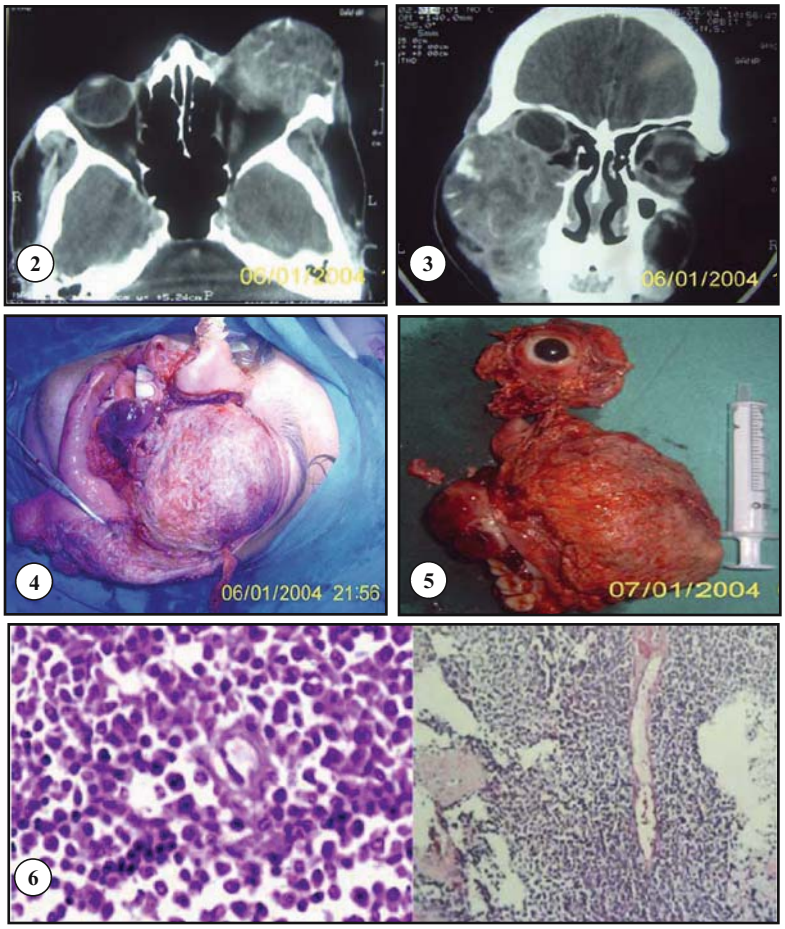
Fig. 2 Axial CT scan

Fig. 3 Coronal CT scan

Fig. 4 Pre-operative photograph

Fig. 5 Excised specimen

Fig. 6 The tumour tissue composed of uniform, small, round or oval cells containing cytoplasmic glycogen and arranged in an peritheliomatous pattern (haematoxylin– eosin x 40, 250)



with progressive painful swelling of the left side of his face for one year. He had epistaxis in small amounts two to three times for 3 months. Nasal obstruction and headache were present for 2 months. Neck had no lymphadenomegaly. He had an operation at a Dental College Hospital 10 months back for some problem in the left side of the face (details were not available). The nasal mass was firm, insensitive and did not bleed on touch, but the facial swelling was red, soft and cystic (Fig. 1). Clinically diagnosed as an antral growth with ethmoidal and orbital involvement. CT examination of the face (5 mm slice thickness, 5 mm intervals, field of view 25cm) was performed. Pre –contrast CT revealed a left paranasal solid mass with intraorbital extension on the axial CT image (Fig. 2). Bone destruction was present on all walls of the sinus cavity. Destruction of the left nasal concha was also noted, with obstruction of the left nasal cavity on the coronal CT images (Fig. 3). The lesion had no abnormal calcification or ossification but the intraorbital portion of the lesion had a cystic area with a sharp margin. The mass showed diffuse homogenous enhancement after intravenous bolus injection of 150 ml iodinated non-ionic contrast medium. Radiology revealed soft tissue swelling in maxilla, with obliteration of nasal fossa and opacification of sinuses with orbital extension. F.N.A.C. from maxillary and nasal mass revealed round cell tumour. After surgical resection of left maxilla with orbital exenteration , using weber fergusson incision (Fig. 4, 5) the facial mass was diagnosed as Ewing's sarcoma histopathologically. Microscopically, the tumour tissue was composed of uniform,small,round or oval cells containing periodic acid Schiff (PAS)- positive cytoplasmic glycogen and sometimes arranged in a "peritheliomatous" pattern (Fig. 6).The tumour cells were immunohistochemically positive for 013 (CD99) but showed negative reaction for keratin, leukocyte common antigen, neuron-specificenolase and synoptophysin. On technetium-99m whole-body scintigraph, no other lesion related to the Ewing's sarcoma was observed. For this reason the patient was accepted as having a primary extraskeletal maxillary Ewing's sarcoma. Post operative chemotherapy was given with VAC protocol (vincristine, actinomycine D, cyclophospamide) and was followed by radiotherapy with a dose of 2000 cGy to the right neck with cervical spinal cord shielding.

Discussion

Ewing's sarcoma represents approximately 4–10% of all bone tumors [17, 18]. Commonly affected regions include: femur (25%), pelvis-ilium (14%), tibia (11%), humerus (10%), fibula (8%), and ribs (6%). When long bones are

affected, Ewing's sarcoma is typically metadiaphyseal in location [17, 18]. Primary Ewing's sarcoma arising from the bones of the head and neck region is extremely rare representing only 1–4% of all Ewing's sarcoma cases [19]. It is the most common malignant bone tumor in children with the peak age being 15 years old. Males are affected twice as much as females (ratio of 2.4:1) [19], and the overwhelming majority of patients are Caucasian (96%) [17, 18].

Primary Ewing's sarcoma of the PNS is usually seen in patients below the age of 20 years, with no gender predilection. The duration of symptoms before presentation ranges from 2 weeks to 2 years. The most common clinical signs and symptoms include severe localized pain, soft tissue mass, fever, leukocytosis, weight loss, and anemia. A painful swelling is the most common clinical presentation [19]. A nasal or submandibular mass might appear later in the course of the disease. Associated anaemia, leukocytosis and fever are common [11]. In cases of intracranial extension, headache, vomiting and papilloedema are common, with localizing signs depending on the site.

Radiologically , Ewing's sarcoma involving long bones presents as a permeative pattern of bone destruction with an aggressive interrupted type of periosteal new bone formation. In the skull these tumours present as permeative, destructive lesions with large associated soft tissue components and no calcification, reflecting the aggressive nature of the tumour. Radiologic features include "Moth-eaten" permeative bony destruction, exuberant periosteal reaction (onion skin, sunburst, spiculated, hair on end), cortical erosion, and presence of an associated soft tissue mass.

CT of Ewing's sarcoma of the PNS shows a diffusely enhancing soft tissue mass with bone destruction [1–5]. Usually no calcification is noted. Similar changes, however, might be seen in the other PNS tumours (squamous cell carcinoma, esthesioneuroblastoma, lymphoma etc.). MRI findings of Ewing's sarcoma of the skull show an unusual pattern of reactive sclerosis [12]. MRI of Ewing's sarcoma typically shows the lesion as hypointense to isointense on T1W1 and hypointense to hyperintense on T2W1, reflecting the pathological finding of densely packed, small round cells with high nuclear : cytoplasmic ratio. Areas of haemorrhage and necrosis demonstrated on MRI are frequent pathological findings, apart from demonstrating increased vascularity. The combination of all of the above findings leads to a strikingly heterogeneous signal within the lesion.

Neither CT nor MRI can give a specific diagnosis but the combination of all of these findings can suggest it. Because of its multiplanar capability, MRI plays a more valuable role in determining lesion extension and facilitating the operative approach. A major advantage of MRI of skull lesions over CT is the lack of streak artefact around thick cortical bone [13]. This also makes MRI more effective in delineating tumour margins.

Ewing's sarcoma is composed of small, round, undifferentiated tumour cells, of uniform appearance, usually crowded in sheets or segregated in lobules by fine fibrovascular septa. Since Ewing's sarcoma are usually vascular, haemorrhagic areas and extensive necrosis are common [11]. Histologically Ewing's sarcoma has a differential diagnosis, in particular from reticulum cell sarcoma of bone,lymphoma, neuroblastoma and olfactory neuroblastoma [23].

The differential diagnosis of masses involving the PNS with a large intracranial soft tissue component and supratentorial extension in all age groups comprises mainly meningioma, [14] metastasis and direct extension from tumours arising below the skull base. Infratemporal masses include paranasal and intracranial extension of nasopharyngeal carcinoma and less often lymphoma and other rare tumours. Diagnosis in these cases is usually made by direct biopsy. Juvenile nasopharyngeal angiofibroma can also extend into the cranial fossa, but the diagnosis of this tumour, arising in adolescent males, is seldom in question owing to its characteristic CT and MRI appearance.

Ewing's sarcoma is a radiosensitive tumour. Multimodality therapy consisting of an initial biopsy, aggressive combination of surgery, chemotherapy and localized radiotherapy is the treatment of choice for Ewing's sarcoma of the head and neck region and may result in long-term survival [19]. Many authors recommend that for treatment of Ewing's sarcoma , the elective therapeutic procedure should consider local irradiation and intermittent but prolonged systemic chemotherapy, leaving mutilating surgery only for recurrent tumours [23]. In general metastatic disease commonly affects the lungs and bones. Pathologic fractures are evident in 2 to 15% of cases. The 5 year survival rate is 60 to 75% [17, 18]. The prognosis of Ewing's sarcoma of the skull, in general, is often good compared with Ewing's sarcoma at other sites. A review of the literature on primary Ewing's sarcoma of the skull revealed evidence of metastases in only one case [15]. Because of the extremely low incidence of metastasis in such cases, long-term survival can be expected. Prognosis is dependent on whether metastases are present at the time of treatment [19]. The 5-year actuarial overall survival (OS) for the treated patients with chemotherapy and radiotherapy was 53%, while the 5-year actuarial disease-free survival (DFS) was 30%. The response to chemotherapy is the only prognostic factor that affects both the OS and DFS [19]. The prognosis of Ewing's sarcoma is improving with radiotherapy and chemotherapy. Further cases are needed to study the biological behaviour of primary cranial Ewing's sarcoma [24].

References

1. Velche-Haag B, Proust F, Laquerriere A, Dehesdin D, Freger P (2002) Ewing's sarcoma of the ethmoid bone:case report. Neurochirugie. 48:25–29
2. Ferlito A (1978) Primary Ewing's sarcoma of the maxilla: a clinico-pathological study of four cases. J Laryngol Otol 92:1007–1024

3. Velche-Haag B, Dehesdin D, Proust F, Marie JP, Andrieu-Guitrancourt J, Laquerriere A (2002) Ewing's sarcoma of the head and neck: a case report. Ann Otolaryngol Chir Cervicofac 119:363–368
4. Csokonai LV, Liktor B, Arato G, Helffrich F (2001) Ewing's sarcoma in the nasal cavity. Otolaryngol Head Neck Surg 125: 665–667
5. Boor A. Jurkovic I, Friedmann I, Plank L, Kocan P (2001) Extraskeletal Ewing's sarcoma of the nose. J Laryngol Otol 115:74–76
6. Howard DJ, Daniels HA (1993) Ewing's sarcoma of the nose. Ear Nose Throat J 72:277–279
7. Lane S. Ironside JW (1990) Extra-skeletal Ewing's sarcoma of the nasal fossa. J Laryngol Otol 104:570–573
8. Amin MN, Islam KM, Ahmed AN, Datta PG, Amin AS, Abdullah M (1990) Ewing's sarcoma of maxilla: a case report. Bangladesh Med Res Counc Bull 16:42–45
9. Pontius KI, Sebek BA (1981) Extraskeletal Ewing's sarcoma arising in the nasal fossa. Light-and electron-microscopic observations. Am J Clin Pathol 75: 410-415.
10. Wexler L, Kacker A, Pior JD, Haddad J Jr. Close LG (2003) Combined modality treatment of Ewing's sarcoma of the maxilla. Head Neck 25:168–172
11. Resnick D, Greenway GG (1996) Tumors and tumor-like lesions of bone: imaging and pathology of specific lesions. In: Bralow L (ed). Bone and joint imaging (2nd edn). Boston, MA: WB Saunders, pp 1058–1097
12. Freeman MP, Currie CM, Gray GF Jr, Kaya JJ (1985) Ewing sarcoma of the skull with an unusual pattern of reactive sclerosis: MR characteristics. J Comput Assist Tomogr 12: 14–36
13. Hudson T, Hamlin DJ, Enneking WF, Pettersson A (1985) Magnetic resonance imaging of bone and soft tissue tumors: early experience in 31 patients compared with computed tomography. Skeletal Radiol 13:134–146
14. Loevner LA, Sonners AL (2002) Imaging of neoplasms of the paranasal sinuses. Magn Reson Imaging Clin N Am 10: 467–493
15. Mishra B, Harass RP, Joseph T, Chandi SM (1993) Primary Ewing's sarcoma of the skull. A report of two cases. Br J Neurosurg 7:683–686
16. M.Harman, F.Kiroglu, M.Kosem ,O Unal (2003) Primary Ewing's sarcoma of the paranasal sinus with intracranial extension: imaging features. Dentomaxillofacial Radiology 32: 343–346
17. Dahnert W (1996) Radiology Review Manual, 3rd Edition, Williams & Wilkins, p 52
18. Resnick D (1996) Diagnosis of Bone and Joint Disorders, 2nd Edition, W. B. Saunders Company, pp 1055–1059
19. Ayman Allam (1999) Ewing's sarcoma of the head and neck: a retrospective analysis of 24 cases. Sarcoma: Taylor & Francis; Volume 3, Number 1 / March 1, 11–15
20. Coskun BU, Cinar U, Savk H, Basak T, Dadas B (2005) Isolated maxillary sinus Ewing's sarcoma. Rhinology. 43(3):225–228
21. Amin MN, Islam KM, Ahmed AN, Datta PG, Amin AS, Abdullah M (1990) Ewing's sarcoma of maxilla--a case report. Bangladesh Med Res Counc Bull 16(1):42–5
22. Fiorillo A, Tranfa F, Canale G, Fariello I, D'Amore R, De Chiara C, Vassallo P, Muto P, De Rosa G, Bonavolonta G (1996) Primary Ewing's sarcoma of the maxilla, a rare and curable localization: report of two new cases, successfully treated by radiotherapy and systemic chemotherapy. Cancer Lett. 103(2):177–82
23. Ferlito A (1978) Primary Ewing's sarcoma of the maxilla: a clinicopathological study of four cases. J Laryngol Otol. 92(11):1007–24
24. Bhatoe HS, Deshpande U (1998) Primary cranial Ewing's sarcoma. British Journal of Neurosurgery. 12(2):165–169
25. Ülkür E. , Açikel, T, Aydin and Çeliköz B (2004) European Journal of Plastic Surgery 27(3):140–142